Guidelines for Diagnosis and Treatment of Myocarditis (JCS 2009)  
– Digest Version –  
JCS Joint Working Group

Revision of the Guidelines

Myocarditis is an inflammatory condition mainly located in the myocardium. Its incidence and mortality in the Japanese population have not been determined, since definitive diagnosis of myocarditis is difficult. These “Guidelines for Diagnosis and Treatment of Myocarditis” have been compiled based on basic and clinical research on myocarditis which has been conducted using sound scientific methods in Japan, though the evidence obtained may not qualify as that of evidence-based medicine. In the guidelines, cardiac magnetic resonance (CMR) imaging is considered valuable as an effective means of diagnosis of myocarditis. In addition, information on cardiac sarcoidosis, autoimmune myocarditis, and drug-induced myocarditis is included.

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I Classification, Diagnosis and Treatment of Acute Myocarditis

1. Etiology

Myocarditis is caused by a variety of bacterial and viral infections. Enteroviruses, especially coxsackievirus B, are often associated with acute myocarditis. However, with the advent of genetic analysis, adenovirus and parvovirus B19 have also been found to be frequent causes of myocarditis.

Exposure to drug treatment, physical stimuli such as radiation and heat, metabolic disorders, immune disorders, and pregnancy are also causes of myocarditis. In the case of idiopathic myocarditis, the etiology is yet to be determined.

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2. Classification

Myocarditis is classified by its etiology, cell type (lymphocytic type, giant cell type, eosinophilic type, granulomatous type), and clinical type (fulminant type, acute type, chronic type) as shown in Table 1.3,4

<table>
<thead>
<tr>
<th>Table 1. Myocarditis Classification</th>
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<tbody>
<tr>
<td>Etiology</td>
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<tr>
<td>Virus</td>
</tr>
<tr>
<td>Bacteria</td>
</tr>
<tr>
<td>Fungi</td>
</tr>
<tr>
<td>Rickettsia</td>
</tr>
<tr>
<td>Spirochetes</td>
</tr>
<tr>
<td>Protozoa, parasites</td>
</tr>
<tr>
<td>Other causes of infection</td>
</tr>
<tr>
<td>Drugs, chemical substances</td>
</tr>
<tr>
<td>Allergy, autoimmune</td>
</tr>
<tr>
<td>Collagen disease, Kawasaki disease</td>
</tr>
<tr>
<td>Sarcoidosis</td>
</tr>
<tr>
<td>Radiation, heat stroke</td>
</tr>
<tr>
<td>Unknown cause, idiopathic</td>
</tr>
</tbody>
</table>

3. Symptoms, Signs and Clinical Tests

1. Symptom

Myocarditis is preceded by flu-like symptoms (chills, fever, headache, muscle aches, general malaise) or gastrointestinal symptoms such as decreased appetite, nausea, vomiting, and diarrhea. Cardiac manifestations of myocarditis appear a few hours to a few days after the initial signs and symptoms.5 Cardiac symptoms consist of (1) those of heart failure, (2) chest pain due to pericardial irritation, and (3) symptoms associated with heart block and arrhythmia. The possibility of myocarditis must be considered if a patient with such symptoms is febrile.

2. Sign

The clinical signs of myocarditis include fever, cardiac rhythm disturbance (tachycardia, bradycardia, and arrhythmia), hypotension, gallop rhythm, rales, jugular venous dilatation, and cardiac tamponade.

3. Blood Biochemistry

Myocarditis is confirmed by the findings of transient elevation of C-reactive protein (CRP), aspartate aminotransferase (AST), lactate dehydrogenase (LDH), the MB form creatine kinase (CK-MB), and cardiac troponin T in blood.6 Troponin T, which can be measured quickly and easily in whole blood by enzyme-linked immunosorbent assay (ELISA), is especially useful for immediate diagnosis. However, which type of troponin, T or I, in more useful for diagnosis has not been determined.7

4. Chest X-Ray

A chest X-ray is useful for visualizing cardiac enlargement and pulmonary congestion.

5. Electrocardiography (ECG)

The ECG is a sensitive and convenient means of diagnosis of myocarditis. It must be timely repeated, since minor abnormalities in the ECG detected initially may become clearer over time. Abnormal ST-T waves and conduction block are frequently observed in myocarditis. A gradual increase in the width of the QRS complex is a sign of exacerbation of myocarditis. Continuous ECG monitoring is crucial to detect potentially fatal arrhythmias.

6. Echocardiography

Myocarditis can be confirmed by transient wall thickening, reduced wall motion and reduced cardiac chamber size in addition to pericardial effusion on echocardiography.8 Echocardiography is especially useful for pediatric patients.

7. Cardiac Magnetic Resonance (CMR)

In addition to the cinematic mode on magnetic resonance imaging (MRI), T1-weighted early signal enhancement and gadolinium-delayed imaging of the heart are useful to make a positive diagnosis of myocarditis.9-11 T2-weighted images reveal the regions of the heart affected by inflammation. CMR imaging can differentiate acute myocarditis from acute myocardial infarction; acute myocarditis exhibits erosive or spotty areas in the epicardium, while lesions of acute myocardial infarction spread from the endocardium like a wave front.

8. Nuclear Medicine Techniques

In gallium-67 (67Ga) myocardial imaging, 67Ga uptake in the myocardium is highly specific for detection of myocarditis, though this method is not highly sensitive.12 On the other hand, technetium-99m (99mTc) pyrophosphate myocardial scintigraphy has relatively high sensitivity but is not specific.13

9. Cardiac Catheterization Including Endomyocardial Biopsy

Cardiac catheterization may be performed in the acute phase of myocarditis if the patient’s condition allows. After coronary lesion has been excluded, an endomyocardial biopsy should be performed to detect myocardial degeneration, myocyte necrosis, inflammatory infiltrates, and/or interstitial edema of the myocardium. Even if the results are negative, the presence of myocarditis cannot be excluded due to the possibility of sampling errors.14 Biopsy sampling at three or more different sites is therefore strongly recommended.15

10. Diagnosis of Viral Infection

Viral infection is confirmed if the viral antibody titer is at least four times higher in an acute phase serum sample than in a sample obtained in remission phase collected at least two weeks apart. However, only approximately 10% of patients with viral infection exhibit a positive antibody titer. Polymerase chain reaction (PCR) is more useful for identifying the genomes of viruses causing myocarditis, but is not commonly performed.16

4. Methods of Diagnosis and Evaluation

Clinical diagnosis of myocarditis should be performed following the “Diagnostic guidelines for acute myocarditis”17 (Table 2).
If acute myocardial infarction is excluded but active lesions of myocarditis can be confirmed on endomyocardial biopsy (Table 3), myocarditis is considered definitively diagnosed.

### 5. Treatment

The primary signs and symptoms and disease progression of myocarditis are relatively easy to grasp. The inflammatory phase lasts one to two weeks, and is followed by a recovery phase. Myocarditis causes myocardial necrosis and inflammation, which result in cardiac dysfunction and failure. Myocarditis is therefore treated in three ways: (1) intervention to eliminate the cause, (2) intervention to improve hemodynamic compromise, and (3) intervention in cardiac dysfunction (Figure 1).

#### 1 Treatment of Asymptomatic or Mildly Symptomatic Myocarditis

Patients with asymptomatic or mildly symptomatic myocarditis with cardiac signs and symptoms should be admitted to the hospital, kept at bed rest, and monitored carefully. A regimen for cardiopulmonary emergency must be prepared beforehand, in the event of acute changes.

#### 2 Treatment of Arrhythmia

Patients with arrhythmia caused by severe heart block should temporarily be treated with external pacing. However, use of drugs must be avoided in case of frequent premature beats and nonsustained in patients with ventricular tachycardia.

#### 3 Management of Heart Failure

For patients with cardiac pump failure in the acute phase, use of catecholamines or carperitide is recommended. If the patient does not respond to treatment, a circulatory assist device should be used.

#### 4 Additional Treatment for Refractory Myocarditis

In patients with persistent inflammation without signs of hemodynamic improvement, short-term treatment with large doses of corticosteroids may be attempted.19 There are cases of...
Case 1: Ventricular tachycardia, ventricular fibrillation, asystole, bystander CPR, minimal complications in central nervous system

CPR

CPR is not successful
Electrical defibrillation (3 to 5 times) for ventricular tachycardia and ventricular fibrillation is ineffective

CPR is successful

Use catecholamines or PDE III inhibitors

Case 2: Low cardiac output and femoral arteriovenous sheath

Use PCPS
In case 1, IABP is concurrently used

1) Initial assisted volume: Start from 3.0 to 3.5 L/min and adjust to the minimum volume which does not cause circulatory failure
2) Place leg bypass from inflow circuit
3) Use anticoagulant: Use ACT 250 sec or 150 to 200 sec if tube is heparin coated. In either case, do not exceed 300 sec.

Maintenance
1) Parameters for circulatory failure: SVO₂, LA, TB, AKBR, acidosis, biochemical tests, urine volume
2) Parameters for cardiac function: Wall motion, EF%, %FS, ejection time, CCI, ETCO₂

Using the above parameters, maintain condition so that cardiac function improves without circulatory failure.

Preparation for device removal
Reduction of assisted volume: Assisted volume is reduced to 0.3 to 0.5 L/min if cardiac function improves. Set assisted volume so ejection time is longest and no circulatory failure occurs. If circulatory failure occurs after volume reduction, return to original volume. The goal is to reduce the volume as much as possible.

Considerations in device removal
Consider device removal if assisted volume is reduced up to 1.5 L/min; parameters for circulatory failure: SVO₂ >60%, TB <3.0 mg/dL, LA normal, no acidosis found on arterial blood gas analysis, no exacerbation of organ dysfunction on biochemical tests, stable urine volume, and parameters for cardiac function: improved wall motion, ejection time >200 msec, ETCO₂, PaCO₂, and CCI >2.0 L/min/m² are observed.

Device removal
Remove the device immediately if no exacerbation of circulatory failure or parameters of cardiac function is observed after assisted volume is reduced to 1.0 L/min.

Measures related to complications of use of devices
1) Multi-organ dysfunction and progressive circulatory failure: Increase assisted flow volume, CVVH, nafamostat mesilate, concurrent use of ulinastatin. Caution for DIC.
2) Leg ischemia: Leg bypass, relaxation incision, amputation
3) Bleeding: Use nafamostat mesilate concurrently, and adjust ACT to 150 to 200 sec. Blood transfusion to maintain Hb 10g/dL, Plt 5.0×10⁴/mm² or more
4) Hemolysis: Administer haptoglobin, and avoid insufficient blood outflow
5) Infection: Determination of pathogen, and administration of antibiotics. Caution for DIC and sepsis.
6) Hyperkalemia: Determination for the cause, eliminate the cause, CVVH, G-I therapy
7) Insufficient blood outflow: Blood transfusion loading targeting PA 20 to 30/10 to 15mmHg

Figure 2. PCPS management of fulminant myocarditis. PCPS, percutaneous cardiopulmonary support; CPR, cardiopulmonary resuscitation; PDE, phosphodiesterase; IABP, intra-aortic balloon pump; ACT, activated coagulation time; SVO₂, mixed venous oxygen saturation; LA, lactic acidosis; TB, tuberculosis; AKBR, arterial ketone body ratio; EF, ejection fraction; FS, fractional shortening; CCI, cerebral circulatory index; ETCO₂, end-tidal carbon dioxide; CVVH, continuous veno-venous hemofiltration; DIC, disseminated intravascular coagulation; Hb, hemoglobin; Plt, platelet count; G-I, glucose-insulin; PA, pression arterielle; PaCO₂, partial pressure of carbon dioxide. Modified from Circ J 2002; 66: 133–144.
remarkable recovery. Treatment with high dose immunoglobulin may also be considered.20

5 Management of Myocarditis in the Recovery Phase
Treatment with angiotensin converting enzyme (ACE) inhibitors and angiotensin receptor blockers can be performed to protect the myocardium.

II Diagnosis and Treatment of Specific Types of Myocarditis

1. Fulminant Myocarditis

1 Background
Fulminant myocarditis causes acute hemodynamic compromise which may prove fatal. External circulatory support is required to save the patient’s life.

2. Diagnosis
The initial evidence of fulminant myocarditis is very similar to that of acute myocarditis, with cardiac shock and circulatory failure as the main crucial points to be handled. On blood chemistry test, blood cardiac troponin is of chief importance for diagnosis.21 A continuous decrease in the cardiac troponin level suggests that the patient is stabilizing. Changes over time in the ECG are important, since widening of the QRS complex and frequent ventricular arrhythmia indicate that myocarditis is following a fulminant course.22 Patients with fulminant myocarditis often have a reduced left ventricular ejection fraction. Monitoring of progressive concentric wall thickening and reduction of wall motion in echocardiography over time is important.8 Fulminant myocarditis can not be identified by histology. Hemodynamics must be continuously evaluated always with echocardiography and guided often by Swan-Ganz catheterization in serious cases.

2. Giant Cell Myocarditis

1 Background
The clinical presentation of giant cell myocarditis tends to be similar to that of fulminant myocarditis.25 Allergy and/or autoimmune factors are thought to be involved in giant cell myocarditis.

3 Treatment
Management of acute fulminant myocarditis should focus on prevention of hemodynamic which have been compromised and bridging the patient to natural recovery.24 Intra-aortic balloon pump (IABP), percutaneous cardiopulmonary support (PCPS), and left ventricular assist device (LVAD) are available as circulatory assistance. Circulatory support should be performed for potentially fatal arrhythmias and low cardiac output. When such devices are used, the following are of great importance: (1) selection of device and timing of introduction, (2) assist volume setting, and (3) prevention of complications4 (Figure 2). In patients refractory to such support, treatment with immunosuppressant may be acceptable. When cardiac dysfunction or heart block does not improve within 3 to 4 days after the initial signs and symptoms of fulminant myocarditis, brief treatment with high-dose corticosteroids or high-dose immunoglobulin may be considered.

Table 4. Diagnostic Guidelines for Eosinophilic Myocarditis

1. Minimally required conditions
1) Increased eosinophil count in peripheral blood (≥500/mm³)a
2) Chest pain, dyspnea, and cardiac symptoms such as palpitations
3) Elevated enzymes indicating myocardial injury, including creatine kinase-MB and the myocardial constitutive protein, including cardiac troponin T
4) ECG changesb
5) Transient left ventricular wall thickeningc and abnormal wall motion on echocardiography

2. Useful information
1) Approximately one-third of patients with eosinophilic myocarditis have allergic conditions (such as bronchial asthma, rhinitis and urticaria).
2) Approximately two-thirds of patients with eosinophilic myocarditis have previous flu-like symptoms (such as fever, sore throat and cough).

3. Endomyocardial biopsy
Histological findings in eosinophilic myocarditis include eosinophil infiltrates, degranulation of eosinophils, disappearance and fusion of cardiomyocytes, and interstitial edema and fibrosis. Occasionally, endocarditis is observed.

a) Some patients have an increased eosinophil count in peripheral blood before cardiac symptoms appear, and some patients have cardiac symptoms with a normal eosinophil count, which gradually increases to above 500/mm³. In the acute phase, the eosinophil count must be determined every 2 to 3 days. However, the eosinophil count increases in a different way in each patient.

b) ST elevation is observed in approximately 50% and abnormal Q waves are observed in approximately one-third of patients with eosinophilic myocarditis. Atrioventricular block, which occurs in viral myocarditis and idiopathic myocarditis, only rarely occurs in eosinophilic myocarditis.

c) Left ventricular wall thickening frequently occurs in eosinophilic myocarditis. Its severity varies among patients. Since wall thickening normalizes within 7 to 14 days, the patient must be monitored over time.

ECG, electrocardiography.

6. Prognosis and Natural History
In the acute phase, myocarditis management of cardiac pump failure and potentially fatal arrhythmias is the main clinical challenge. The prognosis of myocarditis varies depending on the pathogenesis and type of disease.21
3 Eosinophilic Myocarditis

1 Background
Eosinophilic myocarditis is caused by cytotoxic substances such as eosinophilic cationic protein contained in the granules of eosinophils, which infiltrate into the myocardium. Its etiology varies, and includes allergic conditions, drug hypersensitivity, and parasite infection, though it is usually idiopathic.

2 Diagnosis (Table 4)
Eosinophilic myocarditis is diagnosed based on increased eosinophil counts in peripheral blood and significantly increased eosinophil infiltrates, as well as degranulation and degeneration of cardiomyocytes on biopsy. The timing of the onset of increased eosinophil counts in peripheral blood differs among patients.

3 Treatment
Patients with mild eosinophilic myocarditis recover naturally. If the patient has heart failure or serious arrhythmia, corticosteroid treatment is necessary. To prevent cardiac wall thrombi, anticoagulants are used. The prognosis is favorable.

4 Chronic Myocarditis

1 Background
The concept of chronic myocarditis has not been agreed upon between in Japan and outside of Japan. There are reports suggesting the involvement of viral infection or autoimmunity in chronic myocarditis, though the cause of this condition has not been clearly determined.

2 Diagnosis
The diagnosis of chronic myocarditis is difficult. It begins in a latent fashion, but then becomes chronic. Acute myocarditis rarely becomes chronic. The symptoms and signs of chronic myocarditis are non-specific and similar to those of dilated cardiomyopathy. Histological findings are characterized by mononuclear cell infiltrates, aggregated interstitial fibrosis, and fatty infiltration. $^{67}$Ga or $^{99m}$Tc pyrophosphate myocardial scintigraphy and CMR imaging may yield findings suggestive of chronic myocarditis. Blood cardiac troponin level has not been proven to be useful in diagnosis.

3 Treatment
Because the etiology of chronic myocarditis is unclear, palliative treatment such as general treatment for heart failure is performed. It has been reported that chronic myocarditis should be treated based on its pathogenesis, such as viral infection or autoimmunity. However, the effectiveness of immunosuppressant has not been confirmed.

5 Pediatric Myocarditis

1 Background
Pediatric patients with myocarditis include 30 to 40% with the fulminant type and 40 to 50% with the acute type. Chronic myocarditis is rare in pediatric patients. Among pediatric patients, many have viral infections of types seen in daily life. However, adenovirus and enterovirus infections are common causes of pediatric myocarditis. The prognosis of myocarditis in pediatric patients is similar to that in adult patients. The prognosis of fulminant myocarditis is particularly unfavorable in pediatric patients.

2 Diagnosis
The typical manifestation of pediatric myocarditis is elevated blood cardiac troponin. Virus can be detected in feces, urine, blood, and sputum. The findings on ECG and echocardiography in pediatric myocarditis are similar to those in adult myocarditis. Echocardiography is the most useful diagnostic tool for pediatric patients. It must be repeated over time, since the disease may progress rapidly over a few hours in pediatric patients. Nuclear medicine techniques with $^{67}$Ga or $^{99m}$Tc pyrophosphate and CMR may be helpful in diagnosis. An endomyocardial biopsy is relatively safe for older children.

3 Treatment
Once myocarditis is suspected, the patient must be transferred to a facility capable of performing pediatric emergency treatment. The primary goal of treatment is to maintain hemodynamics. Respiratory care and cardiopulmonary circulatory support must be concurrently administered. Treatment with antiviral agents or high dose immunoglobulin should be pathogen-specific.

6 Neonatal Myocarditis

1 Background
Mothers of neonates with myocarditis often have signs of infection a few days prior to delivery, and affected neonates have signs of heart failure before birth. The risk of horizontal vertical infection from the mother to the neonate is high. Two-thirds of neonates with myocarditis have fulminant-type disease, and the mortality rate is high, at 50% or more. Coxsackie B virus infection, which causes fatal myocarditis, accounts for approximately 75% of cases of neonatal myocarditis.

2 Diagnosis
Myocarditis occurs in neonates at birth. Initially, patients are presented with cardiopulmonary signs and symptoms without fever. The signs and symptoms may be non-specific, such as not doing well, feeding difficulty, vomiting, dyspnea, and seizures. Detection of elevated cardiac troponin level is useful for diagnosis. Viruses are isolated in approximately 50% of patients.

3 Treatment
Basically, systemic care and monitoring of the patient is important. Patients must be isolated to prevent spread of infection. Neonates with myocarditis must be immediately transferred to a facility with a neonatal intensive care unit (NICU) or pediatric ICU.
1. Cardiac Sarcoidosis

1 Background
Cardiac sarcoidosis is a systemic granulomatous disease of unknown etiology. The prognosis is closely related to the severity of cardiac manifestations. Attention has been given to Propionibacterium acnes as a cause, since this organism is isolated from tissue affected by sarcoidosis.

2 Diagnosis
According to the guidelines for cardiac sarcoidosis revised in 2006 (Table 5), a myocardial abnormality of unknown cause is occasionally diagnosed as cardiac sarcoidosis after endomyocardial biopsy. If cardiac sarcoidosis is suspected, multidisciplinary collaboration is required for systemic screening for sarcoidosis.

3 Treatment
Corticosteroid treatment is performed for patients with cardiac sarcoidosis, regardless of the severity of cardiac dysfunction. In general, treatment starts at 30 mg/day of prednisolone, and is continued at 5 to 10 mg/day. If arrhythmia or heart failure occurs, the patient should be given standard treatment. Use of a pacemaker, implantable cardioverter defibrillator, drug treatment for heart failure, and cardiac resynchronization therapy should also be considered.

III Diseases Similar to Myocarditis

Table 5. Diagnostic Guidelines for Cardiac Manifestations of Cardiac Sarcoidosis (2006)

<table>
<thead>
<tr>
<th>(1) Patient group diagnosed based on histological findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Histopathological findings include non-necrotizing epithelioid granuloma in the myocardium, and the patient is found to exhibit histopathological changes in organs other than the heart or by clinical signs.</td>
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<table>
<thead>
<tr>
<th>(2) Patient group diagnosed based on clinical signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Histopathological findings do not include non-necrotizing epithelioid granuloma in the myocardium. Patients are diagnosed with cardiac sarcoidosis when they have histopathological changes in organs other than the heart or by clinical signs, together with the following conditions and one or more of six primary diagnostic criteria.</td>
</tr>
</tbody>
</table>

1. Two or more major criteria
2. One major and two or more minor criteria
   1) Major criteria
      (a) Severe atrioventricular block
      (b) Ventricular septal thinning localized at the basal portion
      (c) Abnormal uptake of 67Ga in the heart on 67Ga scintigraphy
      (d) Left ventricular contraction failure (left ventricular ejection fraction less than 50%)
   2) Minor criteria
      (a) Abnormal ECG: Ventricular arrhythmia (ventricular tachycardia, multi-origin or frequent ventricular premature beats), right bundle branch block, axis deviation, or abnormal Q waves
      (b) Echocardiography: Localized abnormal left ventricular wall motion, or morphological abnormalities (ventricular aneurysm and/or ventricular wall thickening)
      (c) Nuclear medicine techniques: Abnormal blood flow on myocardial perfusion scintigraphy (thallium-201 chloride or technetium-99m methoxyisobutylisonitrile, technetium-99m tetrofosmin)
      (d) Abnormal imaging on delayed gadolinium-enhanced cardiac MRI
      (e) Endomyocardial biopsy: Moderate or more severe myocardial interstitial fibrosis and mononuclear cell infiltrates

Primary diagnostic criteria in tests:
1. Bilateral hilar lymphadenopathy
2. Elevated serum angiotensin converting enzyme
3. Negative tuberculin reaction
4. Abnormal uptake of 67Ga in any organ on scintigraphy
5. An increased lymphocyte count and elevated CD4/CD8 ratio in bronchoalveolar lavage fluid
6. Elevated serum or urinary calcium level

Diagnostic exclusion: Giant cell myocarditis must be excluded.

Additional information:
1. Perform coronary angiography to differentiate from ischemic heart diseases.
2. Cardiac manifestations of sarcoidosis occasionally appear a few years after sarcoidosis in organs other than the heart, have been diagnosed. The patient must therefore be followed with ECG and echocardiography on a regular basis.
3. Abnormal uptake of fluorine-18 fluorodeoxyglucose PET in the heart is a useful diagnostic clue in cardiac sarcoidosis.
4. Some cases of cardiac sarcoidosis are manifested only by complete atrioventricular block without any other minor criteria as listed.
5. Cardiac sarcoidosis may initially manifest itself as pericarditis (as shown by ST segment elevation or pericardial effusion in the ECG).
6. Cases of non-necrotizing epithelioid granuloma are not observed frequently on the endomyocardial biopsy.

67Ga, gallium-67; MRI, magnetic resonance imaging; ECG, electrocardiography; PET, positron emission tomography. Adapted from The Japanese Journal of Sarcoidosis and Other Granulomatous Disorders 2007; 27: 89–102.
2. Autoimmune Myocarditis (Myocarditis Derived From Collagen Diseases)

1 Background
Initial manifestations of autoimmune myocarditis include dysfunction of the kidneys, skin, choroid plexus, and inflammation not involving infection such as deposition of immune complexes and activation of complement.

2 Diagnosis
Initially, autoimmune myocarditis rarely occurs with myocarditis alone. Pericarditis is associated with the severity of disease activity. A presence of antinuclear antibodies in pericardial effusion, autoimmune bodies, reduced complement activity, and elevated immune complex levels are supportive of the diagnosis. Echocardiography, myocardial scintigraphy and CMR imaging are used for diagnosis, but an ordinary endomyocardial biopsy is not diagnostic. Scleroderma, systemic lupus erythematosus, polyynoisitis, dermatomyositis, rheumatoid arthritis, polyarteritis nodosa, and allergic granulomatous angitis (Churg-Strauss syndrome) are associated with cardiac manifestations.

3 Treatment
Autoimmune myocarditis is treated with corticosteroids or concurrent use of immunosuppressant if the patient has decreased cardiac function, severe pericardial effusion, or concurrent dysfunction of other organs.

IV Conclusion
The diagnosis of myocarditis is difficult. The first step in diagnosis is to suspect myocarditis. The primary principles of treatment are to make the clinical diagnosis and manage cardiopulmonary emergency promptly. Every effort must be made to confirm the diagnosis of myocarditis by histology, since some cases of specific myocarditis may respond to corticosteroid treatment.

References
15. Cooper LT, Baughman KL, Feldman AM, Frustaci A, Jessup M, Kuhl U, et al. The role of endomyocardial biopsy in the management of cardiovascular disease: A scientific statement from the American Heart Association, the American College of Cardiology, and the European Society of Cardiology. Endorsed by the Heart Failure


**Appendix**

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